

CASE REPORT



I can't hear you, you said I had what?: A case report and literature review

Zackquill J. Morgan, Leslie Cler and Leigh Hunter

Internal Medicine, Methodist Dallas Medical Center, Dallas, TX, USA

ABSTRACT

We report the case of a 46-year old African American woman who presented to the emergency department with one week of progressive bilateral deafness associated with worsening gait abnormalities, visual changes, and confusion. She was diagnosed with Wernicke encephalopathy (WE) attributed to alcohol abuse; her symptoms, including hearing loss, improved with thiamine replacement. WE, a condition due to thiamine deficiency, commonly affects those with alcohol use disorder or gastric bypass history. Though traditionally associated with a triad of encephalopathy, ophthalmoplegia, and ataxia, it can be more rarely associated with auditory deficits or other neurologic findings. Though hearing loss has previously been reported as a rare symptom of WE, it has not been described in WE due to alcohol abuse. We performed a review of the literature to determine if WE associated with hearing loss had been previously reported.

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1. Case description

A 46 year old African American woman presented with one week of worsening hearing loss along with three days of vision changes, confusion, visual hallucinations, and difficulty ambulating. She had a history of invasive ductal cell carcinoma status-post mastectomy and chemotherapy. Additional history included hemorrhoids with recent hemorrhoidectomy necessitated by excessive bleeding while on chemotherapy.

Physical examination was notable for a reasonably well-nourished woman, although her BMI had dropped from 27 to 22 within the previous 18 months. Vital signs were within normal limits other than tachycardia with a heart rate up to 110 beats per minute. She was alert, intermittently oriented, and complained of visual hallucinations. She also spoke very loudly and complained of difficulty hearing verbal questions or reading written questions. Responses were inappropriate at times, which could have been attributable to misunderstanding questions asked. Neurologic examination was significant for end-gaze horizontal nystagmus, dysmetria as measured by finger-to-nose exam, decreased visual acuity and decreased hearing bilaterally. She had normal sensation, motor strength, proprioception, and reflexes. Her gait was unsteady, and she exhibited dysdiadochokinesis. Laboratory evaluation was significant for white blood cell count (14.4×10^3 u/L), aspartate aminotransferase (106 U/L), and alanine aminotransferase (15 U/L). The patient

was initiated on broad spectrum antimicrobials for suspicion of meningoencephalitis. Computed tomography (CT) and magnetic resonance imaging (MRI) scans of the brain were normal and lumbar puncture findings were also normal with negative cultures and polymerase chain reaction results, thus antimicrobials were stopped. Wernicke encephalopathy (WE) was suspected, and intravenous (IV) thiamine supplementation was initiated. Auditory and visual disturbances, confusion, and ataxia subsequently improved. Serum thiamine levels returned undetectable and she later admitted to significantly more alcohol intake than the 'occasional use' reported at time of admission. Her hearing returned to baseline as did her mental status and vision. She continued to have somewhat increased difficulty ambulating at the time of discharge.

2. Discussion

WE classically presents as a triad of encephalopathy, oculomotor dysfunction, and ataxia; only one-third of patients exhibit the full triad [1]. Other less common findings include vestibular dysfunction, autonomic dysregulation, peripheral neuropathy, hypothermia, protein calorie malnutrition, and hearing loss [2]. While traditionally associated with alcohol intake, WE can also occur with gastric bypass, inflammatory bowel disease, prolonged artificial nutrition, or other disease states with high metabolic demand [3].

Thiamine is a key co-factor in the metabolism of glucose, the primary fuel of brain tissue. In the absence of this co-factor, continued glucose metabolism generates free radicals [3] and can precipitate NMDA (N-methyl-D-aspartate) receptor-mediated excitotoxicity [4]. Thiamine deficiency in alcoholism is due to a combination of decreased intake and reduced absorption [3]. Diagnosis is based solely on clinical findings, although laboratory or imaging data can help support the diagnosis. MRI findings, when present, typically reveal symmetric FLAIR (fluid-attenuated inversion recovery) hyperintensities with contrast enhancement in the midbrain. MRI has a specificity of 93% with a sensitivity of 53% in WE diagnosis [5,6]. Treatment is initially IV thiamine repletion, followed by oral thiamine repletion which should be continued as long as the patient remains at risk for deficiency. Residual deficits are common, with nystagmus being the most common residual symptom followed by ataxia [4]. IV thiamine should be given prior to glucose-containing substances to avoid precipitating Korsakoff syndrome – a syndrome of retrograde and anterograde amnesia. It is imperative that physicians consider this entity in the presence of reported alcohol intake even when classic findings are not present. Increased recognition of this disease should facilitate prompt treatment and help reduce morbidity and mortality.

In a paper by Walker and colleagues, eight unique cases of WE associated with hearing loss were identified [7]. Further review of English-language literature found two additional cases of hearing loss associated with WE. The patients and their findings are summarized in Table 1. Of the previously reported cases, one was associated with hyperemesis gravidarum [8], one was associated with Crohn's disease and colectomy [9], five were associated with bariatric surgery (7, 10, 11, 12 and 13), one was attributed to a strictly limited diet following acute pancreatitis [14], one was associated with parenteral nutrition and chemotherapy for gastric carcinoma [15], and one was attributed to gastroparesis associated with diabetes mellitus [7], and Our patient represents the eleventh patient described in accessible literature. The current case stands out for a number of reasons. First, this is the only case in which alcohol abuse is theorized to be the primary etiology of thiamine deficiency manifesting as hearing loss; all other described cases were attributable to a GI source. This finding may suggest that the mechanism or relative speed of thiamine depletion may confer different patterns of neuronal damage, producing auditory symptoms more often when thiamine is depleted through means other than alcohol abuse. Second, this case is unique as it is the only case described without MRI changes (no MRI results were reported for patient 7 [15]). All other described cases demonstrated MRI changes

Table 1. A summary of published cases of hearing loss associated with Wernicke encephalopathy.

Patient No.	Patient Age/Sex	Etiology of Thiamine Deficiency	Auditory Symptoms	MRI Findings	Outcome	Reference
1	17 F	Hyperemesis gravidarum	Bilateral hearing loss	Bilateral thalamic hyperintensities	Improved with thiamine repletion	[8]
2	31 F	Crohn's disease	Bilateral tinnitus	Bilateral inferior colliculi abnormalities	Resolved after 48 hours of IV thiamine repletion	[9]
3	35 F	Bariatric surgery	Bilateral hearing loss	Bilateral symmetric, enhancement of the periaqueductal grey matter, medial thalami, premotor and motor cortices	Not reported	[10]
4	44 F	Bariatric surgery	Bilateral hearing loss	Abnormal symmetric intraparenchymal signal of the bilateral thalami	Hearing loss improved, but still present 24 hours after IV thiamine repletion	[7]
5	35 F	Bariatric surgery	Bilateral hearing loss	Periaqueductal grey matter and medial thalamic nuclei hyperintensity	Not reported	[11]
6	23 M	Strictly limited diet	Bilateral hearing loss	Hyperintensity of the bilateral inferior cerebellar peduncles, thalamus and mammillary bodies	Hearing loss resolved within 48 hours of IV thiamine therapy	[14]
7	28 F	Gastric carcinoma treated with chemotherapy	Bilateral deafness	Not reported	Hearing loss resolved after IV thiamine therapy	[15]
8	61 F	Diabetic gastroparesis	Bilateral hearing loss	Periaqueductal grey matter, bilateral superior colliculi, mammillary bodies and fornices, hypothalamic region, and both medial thalami	Hearing loss resolved within 24 hours of treatment	[7]
9	35 F	Bariatric surgery	Bilateral hearing loss	FLAIR hyperintensities of the bilateral mammillary bodies and periaqueductal grey matter	Hearing improved, but patient remained quadriplegic	[12]
10	27 M	Bariatric surgery	Bilateral hearing loss	T2 hyperintensities of the bilateral posteromedial thalamus, the hypothalamus and the bilateral maxillary bodies	Hearing loss resolved within 2 weeks of treatment	[13]
11	46 F	Alcohol	Bilateral deafness	No MRI changes	Hearing loss resolved with treatment	Current study

typical for WE despite a reported sensitivity of only around 50%. This again raises the possibility of separate pathophysiologic mechanisms by which neuronal injury occurs in WE, resulting in more common changes on MRI of the brain. Third, our patient stands out as the second oldest patient described to develop hearing loss as part of the constellation of symptoms. Only patients 4 and 8 (ages 44 and 61, respectively) were older than 35 with our patient being 46. This suggests that there might be different age-related mechanisms behind neuronal injury in WE. A mechanism that perhaps may deteriorate with age making auditory symptoms less common in older populations.

3. Conclusion

Physicians must consider WE in any patient with some (or all) of the symptoms of the classic WE triad: ophthalmoplegia, ataxia, and confusion. Additional care should be taken to avoid dextrose administration prior to repletion of thiamine in these patients. Without reliable biomarkers or imaging findings, clinical diagnosis will remain the mainstay of WE diagnosis. More research is needed to understand the pathophysiology of WE and how manifestations relate to various mechanisms of thiamine deficiency. Hearing loss is a rare, but documented symptom of WE. Hearing loss may be more prevalent in younger populations with GI tract-associated thiamine deficiency, but also may present due to alcoholism.

Disclosure statement

No potential conflict of interest was reported by the authors.

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